International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Marging Y et al. Int J Reprod Contracept Obstet Gynecol. 2025 Mar;14(3):951-953
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20250539

Case Report

Successful pregnancy and delivery in a rare case of pemphigus vulgaris

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Received: 03 January 2025
Accepted: 04 February 2025

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ABSTRACT

Pemphigus vulgaris (PV) is a rare autoimmune bullous dermatosis characterized by flaccid blisters and extensive erosions on the skin and mucous membranes. While in the active stage of PV lesions, it is reported to be linked to infertility, and its presence during pregnancy is exceedingly rare. Pregnancies complicated by pemphigus vulgaris often result in adverse obstetrical outcomes, including prematurity, neonatal skin lesions, and even fetal demise. Our patient, a chronic pemphigus vulgaris case, was being treated with rituximab, a pregnancy category C drug. Later upon conceiving, her medication was switched to oral steroids, which were later changed to intravenous steroids due to a flare-up in the third trimester. Her elective lower segment caesarean section was done with delivery of a female baby weighing 2.6 kg with APGAR scores of 8 and 9 at one minute and five minutes respectively, without any apparent skin lesions. During the postpartum period, her skin lesions resolved completely by the time of her routine postnatal follow-up.

Keywords: Pemphigus vulgaris, Rare, Autoimmune, Bullous dermatosis, Steroids, Pregnancy, Neonatal outcome

INTRODUCTION

Pemphigus vulgaris (PV) is exceedingly an uncommon immune-mediated bullous disorder characterized by aggressively erosive vesicular and bullous blisters, commonly affecting the skin and mucosa of any part whole body like trunk, extremities, face, oral cavity even scalp.[1] Genetic factors has a significant part in the predisposition to PV, as first-degree relatives are more likely to develop autoimmune diseases.[2]

PV is rare in the general population and even more so during pregnancy. Women with PV might face infertility issues, potentially related to the chronic nature of the disease, the medications used for treatment, or the presence of other autoimmune disorders. Immunosuppressive therapies commonly used in PV can affect reproductive health and obstetric outcomes in terms of preterm deliveries, intrauterine foetal demise, stillbirths and baby born with autoimmune diseases.[3] The pathogenesis of PV is due the formation of autoantibodies which act against desmosomes which are the transmembrane glycoproteins, specifically desmoglein 3 (Dsg3) a protein crucial for cell-to-cell adhesion in the epidermis. This leads to the depletion of Dsg3 in desmosomes resulting into the disruptions of the integrity of the desmosomes and ultimately leads into the depletion of cell-to-cell adhesion between keratinocytes (skin cells) in the basal as well as in the suprabasal layers of the epidermis. This process, known as acantholysis, causes the separation of keratinocytes as a result of which there is formation of clefts and blistering intraepidermally. The diagnosis of pemphigus vulgaris usually confirmed through skin biopsy and direct immunofluorescence.

Histologically, the condition is marked by acantholysis and suprabasal cleft formation, often described as resembling a "row of tombstones". Direct immunofluorescence typically reveals the presence of IgG antibodies against the PV antigen throughout the epidermis.[4]

CASE REPORT

30-year-old primigravida (G1P1) married for two years at 32 weeks and 3 days by her last menstrual period (LMP) presented to the dermatology department on 24/3/24. She had been a follow-up case for chronic pemphigus vulgaris, diagnosed in 2019, and was treated with two doses of rituximab and DCP pulses, achieving remission until 2022. She is also HIV positive and has been on antiretroviral therapy (ART) for the past year. Her family history is notable for a younger sister who had a similar condition and passed away in 2010.Recently, the patient experienced an exacerbation of PV, with multiple fluid-filled blisters, active pus discharge, and crusting on the soles, palms, back, abdomen, and limbs (Figure 1a and b) over the past week, accompanied by oral candidiasis. The lesions were painful, itchy, and caused a burning sensation, and oral lesions made swallowing difficult, though she had no breathing difficulties.

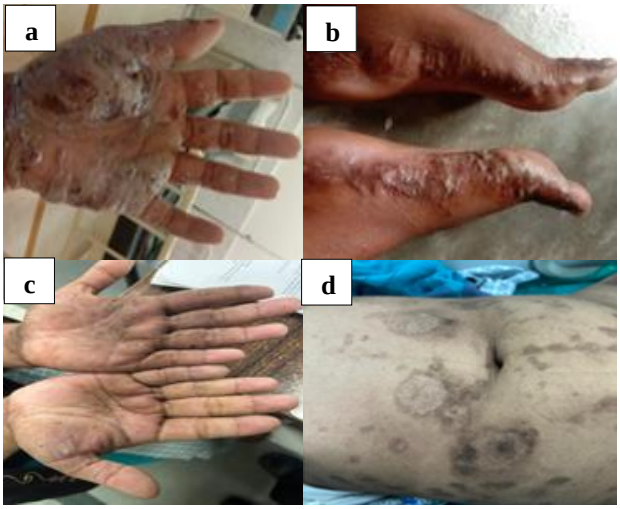

Figure 1: (a and b) Showing multiple erosive vesicles and blisterings in the flare up stage, and (c and d) showing the lesions being dried and crusted healed after corticosteroids).

Histopathology of skin biopsy from left thigh showed eosinophils, neutrophils and acantholytic cells in epidermis and perivascular mild inflammatory infiltrates in dermis (Figure 2a and b) with immunofluorescence IgG:3+ intercellular pattern and C3:2+ intercellular pattern. Initial treatment included intravenous ceftriaxone, later switched to piperacillin-tazobactam based on culture and sensitivity reports, along with clotrimazole mouth paint for the oral candidiasis. Consultation with the ART center was done and under the continous supervisions of the obstetrics and gynecology team intravenous dexamethasone at 1.5 cc (6 mg) once daily was started. As her condition improved, the dose was tapered to 1 cc (4 mg). She received regular antenatal check-ups and vigilant fetal surveillance was done. An ultrasound confirmed a healthy pregnancy, and she underwent an elective lower segment cesarean section (LSCS) at 37 weeks of gestation on 26 April 2024, delivering a female baby weighing 2.6 kg, cried immediately after birth with good APGAR score. The LSCS scar were healthy and stitches were completely removed on 06 May 2024. At the time of discharge, the patient was hemodynamically stable and advised to return for a follow-up in one week with tab. Prednisolone 10 mg in tapering doses. At her routine postnatal follow-up, the patient's lesions had almost completely healed and dried up, with no new lesions observed (Figure 1c and d). Baby was also healthy without any skin lesions particularly.

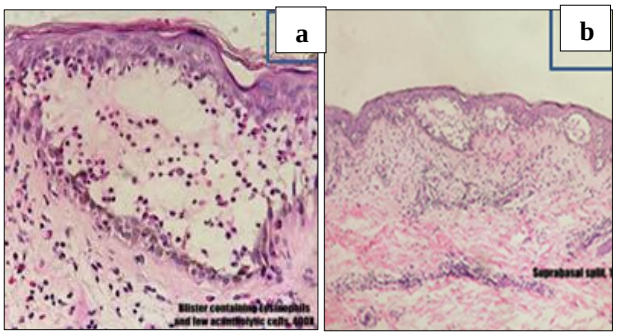

Figure 2: Histopathology of skin blisters, (a) showing blister containing eosinophils acantholytic cells at 400X, and (b) demonstrating suprabasal cleft formation a 100X, often described as resembling a "row of tombstones".

DISCUSSION

PV is a group of rare potentially fatal autoimmune bullous and erosive dermatosis characterized by widespread blisterings, bullae, and erosions on the skin and mucosal membranes.[5] Pregnancy with PV is particularly rare, primarily due to the disease's association with infertility during active phases.[6] Recently According to the English literature published on PubMed wherein they have reviewed all the reported cases of PV and pregnancy found out that the reported stillbirth rate to be 10% (5 out of 49 patients), with a rate of 12% (6 cases) perinatal mortality.[7] The interplay between PV and pregnancy complicates treatment for clinicians. The primary treatment for PV during pregnancy involves glucocorticoids, which remain the cornerstone of therapy both during and outside of pregnancy.[8] Currently, rituximab which is a chimeric anti-CD20 monoclonal antibody acting against the B cells, has shown significant success in the management of Pemphigus vulgaris and it has a steroid-sparing property too. However, rituximab is contraindicated during pregnancy because of the fact that it depletes the fetal B lymphocytes depletion.[9]

Management of delivery in PV patients requires careful consideration. We should be cautious while conducting vaginal delivery as any form of trauma to the vagina during childbirth can lead to the spreading and deterioration of PV lesions. Therefore, the choice between vaginal delivery and cesarean section should be carefully evaluated on the basis of patient's condition and the potential risks involved.[10]

CONCLUSION

PV during pregnancy is a complex and challenging scenario. Despite the associated infertility during active phases of the disease, successful pregnancies can occur with meticulous management and a multidisciplinary approach. Continued research and case documentation are essential to further understand and improve outcomes for pregnant women with PV.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Kridin K. Pemphigus group: Overview, epidemiology, mortality, and comorbidities. Immunol Res. 2018;66(2):255-70.
2. Firooz A, Mazhar A, Ahmed AR. Prevalence of autoimmune diseases in the family members of patients with pemphigus vulgaris. J Am Acad Dermatol. 1994;31:434-7.
3. Kardos M, Levine D, Gurcan H, Ahmed RA. Pemphigus vulgaris in pregnancy: analysis of current data on the management and outcomes. Obstet Gynecol Surv. 2009;64:739-49.
4. Joly P, Gilbert D, Thomine E, Zitouni M, Ghohestani R, Delpech A, et al. Identification of a new antibody population directed against a desmosomal plaque antigen in Pemphigus and Pemphigus foliaceus. J Invest Dermatol. 1997;108:469-75.
5. Fainaru O, Mashiach R, Kupferminc M, Shenhav M, Pauzner D, Lessing JB. Pemphigus vulgaris in pregnancy: A case report and review of literature. Hum Reprod. 2000;15(5):1195-7.
6. Kardos M, Levine D, Gurcan H, Ahmed RA. Pemphigus vulgaris in pregnancy: analysis of current data on the management and outcomes. Obstet Gynecol Surv. 2009;64:739-49.
7. Kardos M, Levine D, Gürcan HM, Ahmed RA. Pemphigus vulgaris in pregnancy: Analysis of current data on the management and outcomes. Obstet Gynecol Surv. 2009;64:739-49.
8. Ramsey-Goldman R, Schilling E. Immunosuppressive drug use during pregnancy. Rheum Dis Clin North Am. 1997;23:149-67.
9. Genovese G, Derlino F, Berti E, Marzano AV. Treatment of autoimmune bullous diseases during pregnancy and lactation: A review focusing on pemphigus and pemphigoid gestationis. Front Pharmacol. 2020;11:583354.
10. Goldberg NS, DeFeo C, Kirshenbaum N. Pemphigus vulgaris and pregnancy: Risk factors and recommendations. J Am Acad Dermatol. 1993;28(5):877-9.

Cite this article as: Marging Y, Sharma N, Verma S, Singh AS. Successful pregnancy and delivery in a rare case of pemphigus vulgaris. Int J Reprod Contracept Obstet Gynecol 2025;14:951-3.